WEST SYNDROME AND INFANTILE SPASMS: FAQs

What are infantile spasms?

Infantile spasms are a type of seizures that tend to affect children under the age of 2 years. In this, children have tightening and jerky movements of limbs and body. These usually occur in clusters. Clusters of spasms usually occur upon awakening but can happen at any time of the day.

What is West Syndrome?

West syndrome is a clinical condition characterised by the combination of

- Infantile spasms
- Abnormal EEG showing chaotic pattern called Hypsarrhythmia
- Delayed development or loss of acquired milestones

Why is it called 'West' Syndrome?

West Syndrome is named after the English surgeon Dr.William James West, who published the first description of infantile spasms in 1841, having witnessed the disease in his own son, James E West. He named the seizures "Salaam Tics" at the time.

How do I recognise infantile spasms?

Infantile spasms manifest in different forms. These usually consist of a brief interruption of behaviour, with lifting and extension of the arms and bending forward at the waist. This may be accompanied by a rapid and forceful drop of the head. The individual spasms last a few seconds. Spasms often occur in clusters and the child may have several of them in a row**,** although singles spasms are possible. After the first spasm, there is a pause and then another spasm follows. Each cluster typically lasts a few minutes. After individual spasms, children often cry. They usually occur as child wakes up from sleep.

What is the cause of infantile spasms?

Almost any brain abnormality or injury to the brain has the potential to cause infantile spasms. Examples include brain malformations, brain infections, problems at birth and neonatal period such as delayed cry leading to lack of oxygen or low sugar levels.

About one-tenth of children with infantile spasms may not have an identifiable cause despite detailed evaluation.

What is the outcome of West syndrome?

The outcome of West syndrome is highly variable; however, it is generally difficult to treat and commonly associated with developmental delay and future epilepsy. Early diagnosis and treatment of West syndrome helps in better seizure control and improved developmental outcome.

How is West Syndrome diagnosed?

The main modality of diagnosis of West Syndrome is an electroencephalogram (EEG), on which a chaotic pattern of brain waves called as hypsarrhythmia is the characteristic abnormality noted in West Syndrome. Apart from this, other investigations like MRI of the brain are done to find out the underlying cause.

How is West Syndrome treated?

A number of treatment modalities are available. The most effective drug therapies include ACTH and steroids and a drug called *vigabatrin*. Other anti-seizure medications are also used, though these are less effective. These include valproate, zonisamide, topiramate , clonazepam and clobazam. A one-week trial of pyridoxine is usually given at the beginning in all patients.

What are the side effects of steroids?

High dose steroids used for treatment of West Syndrome. Prolonged steroid therapy can have side effects like

- Irritability
- Increased appetite
- Weight gain
- High blood pressure
- High blood sugar levels
- Increased risk of infections.

There are however transient and improve when steroid therapy is stopped.

What precautions need to be undertaken while the child is on steroids?

Any vaccines should be given to the child after consultation with the treating doctor. Children on high dose steroids for more than 2 weeks must not be given live vaccines like oral polio vaccine, measles, mumps, varicella vaccines until at least 1 month of stopping the steroids. These children are more prone to infections; hence parents should report to the hospital if the child has fever.

What are the side effects of other drugs?

Vigabatrin can cause visual disturbances when used for long duration. Valproate can cause affect liver and pancreas in high doses.

Is there any treatment apart from drugs?

In children who do not respond to anti-seizure or hormonal therapies, other modalities like surgery (to remove seizure-generating parts of the brain) and ketogenic diet may be tried.

What is ketogenic diet?

Ketogenic diet is a specialized very-high fat diet used in treating many types of epilepsy. It involves strict adherence to a diet plan and required monitoring by an experienced neurologist.

How long is the treatment continued?

Steroids are usually given for about 2 weeks followed by slowly tapering the dose and stopping. Vigabatrin is usually continued for 6 months. Other anti-seizure medications are usually continued for a couple of years till seizure control is attained.